acta medica

LETTER TO THE EDITOR

A Case of Pneumatosis Intestinalis Associated with Sjogren's Syndrome

Seiichi Horinaka[1]
ORCID: 0000-0003-0476-1504

Masashi Ohe[1]
ORCID: 0000-0002-6684-6688

Haruki Shida[1]
ORCID: 0000-0001-6331-0952

Ken Furuya[1]
ORCID: 0000-0003-3373-4337

Misturu Sugiura[2]
ORCID: 0000-0001-6972-7070

[1]Department of Internal Medicine, JCHO Hokkaido Hospital, Sapporo, Japan

[2]Department of Diagnostic Radiology, JCHO Hokkaido Hospital, Sapporo, Japan

Corresponding Author: Masashi Ohe
Department of Internal Medicine, JCHO Hokkaido Hospital, Sapporo, Japan.
E-mail: oektsp1218@sweet.ocn.ne.jp

Received: 3 January 2022, Accepted: 18 March 2022, Published online: 13 June 2022

A 68-year-old female patient with primary Sjogren's syndrome (SjS) was referred to our hospital due to dry cough exacerbation and abdominal fullness. However, she complained of neither abdominal pain nor constipation. She was diagnosed with SjS accompanied by interstitial pneumonia based on sicca symptoms, positive Schirmer's test, elevated levels of anti-SSA antibody, positive sialography, and honeycomb opacities on chest computed tomography (CT), 7 years ago, and was successfully treated with corticosteroids (CSs), such as prednisolone (PSL) and methyl PSL. During the last 4 years, the patient received methyl PSL at 4 mg/day. On this visit, the clinical presentation showed a distended abdomen and weak bowel sounds, without rebound tenderness. Laboratory findings were as follows: white blood cell (WBC) count of 5,610/μL (of which neutrophil cells were 68.7%); hemoglobin (Hb) level of 11.5 g/dL; platelet (Plt) count of 25.3 × 10^4/μL; C-reactive protein level (CRP) of 0.20 mg/dL; and lactate dehydrogenase (LDH) level of 170 U/L (Table 1). Chest CT scan revealed honeycomb opacities in both lower lung fields, and intra-abdominal free air indicating pneumoperitoneum (red arrow) and dilated intestinal canals, wherein the walls contain air-filled cysts (Figure 1A). Abdominal CT scan also revealed pneumoperitoneum, and dilated intestinal canals, wherein the walls contain air-filled cysts (Figure 1B, C). Based on these findings, the patient was diagnosed as having a complication of pneumatosis intestinalis (PI) and not gastrointestinal perforation. The patient was advised to eat very slowly and prohibit the habit of drinking carbonated water. She was treated with dextromethorphan hydrobromide hydrate for deteriorating dry cough, mosapride citrate hydrate for intestinal hypomotility, and dimethicone for abdominal fullness. However, the aforementioned symptoms did not improve within 3 months after treatments. Furthermore, the abdominal CT findings were exacerbated (Figure 2A, B). Therefore, taking notice of constipation, she received an add-on dihydrocodeine phosphate treatment. Three months thereafter, the aforementioned symptoms and abdominal CT findings considerably improved (Figure 3A, B).

In the present case, PI with pneumoperitoneum mimicked gastrointestinal perforation; however, there was no clear evidence of gastrointestinal perforation and peritonitis because the patient had no abdominal pain and rebound tenderness, and there was no evidence of peritoneal inflammation in laboratory data.

Table 1. Laboratory data

WBC	5,610/μL	TP	6.8 g/dL
Baso μ	0.4 %	Alb	3.7 g/dL
Eosino	1.8%	Na	142 mEq/L
Neutro	68.4%	Cl	108 mEq/L
Lympho	20.7%	K	4.2 mEq/L
Mono	8.4%	Ca	9.1 mg/dL
RBC	383x104 /μL	BUN	11.2 mg/dL
Hb	11.5 mg/dL	Cr	0.38 mg/dL
Plt	25.3x104 /μL	UA	4.0 mg/dL
		Fe	79 mg/dL
TB	0.5 U/L	TG	174 mg/dL
CPK	37 U/L	LDL-C	84 mg/dL
LDH	170 U/L		
ALP	81 U/L	CRP	0.2 mg/dL
GOT	21 U/L		
GTP	13 U/L	FBS	85 mg/dL
γGTP	21 U/L	HbA1c	5.5%

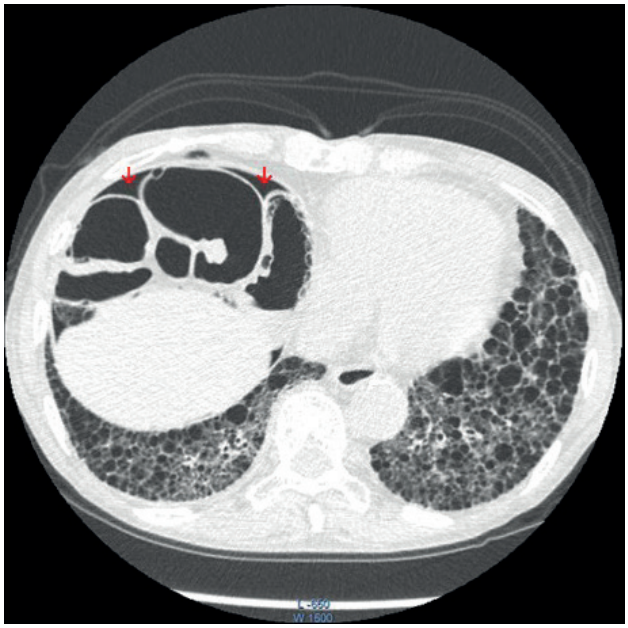
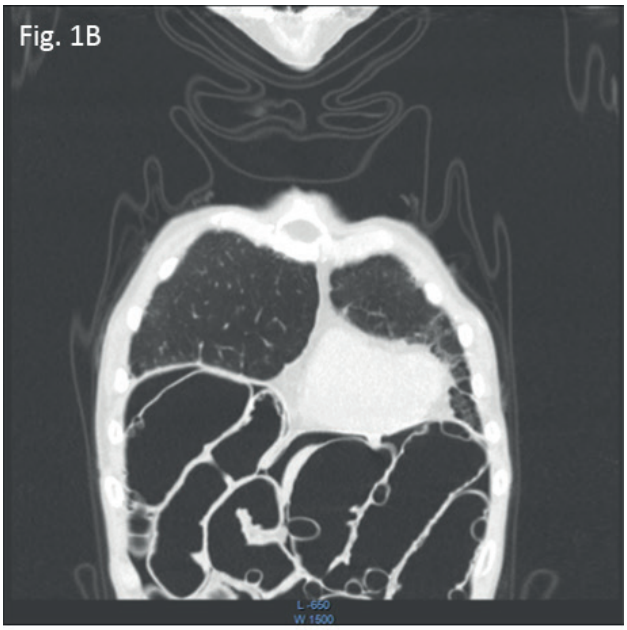


Figure 1A. Chest CT scan reveals honeycomb opacities in both lower lung fields, intra-abdominal free air indicating pneumoperitoneum (red arrow), and dilated intestinal canals, wherein the walls contain air-filled cysts.

PI is a rare disease characterized by the presence of gas in the gastrointestinal wall and is known to be associated with several clinical conditions, such as pulmonary diseases (e.g., asthma, cystic fibrosis and COPD), gastrointestinal diseases, and traumatic injury, as well as connective tissue diseases [1,2].



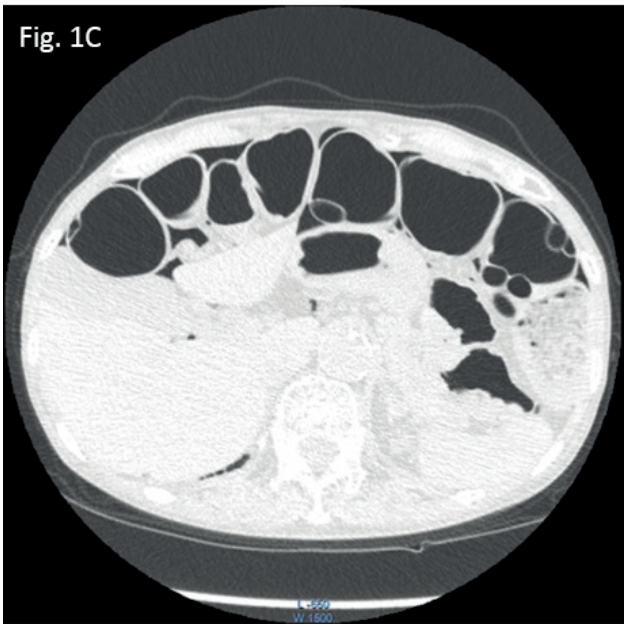


Figure 1B, C. Abdominal CT scan reveals pneumoperitoneum and dilated intestinal canals, wherein the walls contain air-filled cysts.

Especially, PI is commonly seen in systemic sclerosis (SSc) but rarely in systemic lupus erythematosus (SLE) [3]. Similarly, few cases of PI associated with SjS have been reported [3-6]. In patients with SSc, smooth muscle cell atrophy and fibrosis in the gastrointestinal walls are observed, which lead to intestinal hypomotility. Bacterial overgrowth and bowel distention are observed in this condition, which can lead to elevated intraluminal pressure and force gas into the intestinal walls [1]. On the other hand, intestinal vasculitis is speculated to be associated with PI in patients with SLE [7]. Contrarily, etiologies of PI associated with SjS remain

unclear [3]. CSs are reported to be associated with PI. CSs decrease lymphatic tissue in the intestinal Peyer's patch cells, resulting in degeneration of the mucosa that leads to the entry of gas into the peritoneum and intestinal wall [3]. As mentioned above, pulmonary disease is a cause of PI. A severe cough can trigger alveolar rupture, which can result in the introduction of air along the vascular channels in the mediastinum, tracking down to the retroperitoneum, and then to the mesentery of the bowel [1]. Consequently, severe cough is thought to be associated with PI.

In the present case, not only the long-term administration of CSs but also exacerbated cough due to interstitial pneumonia associated with SjS were thought to be one of the etiologies of PI since dihydrocodeine phosphate improved PI. Previous reports regarding PI associated with SjS revealed

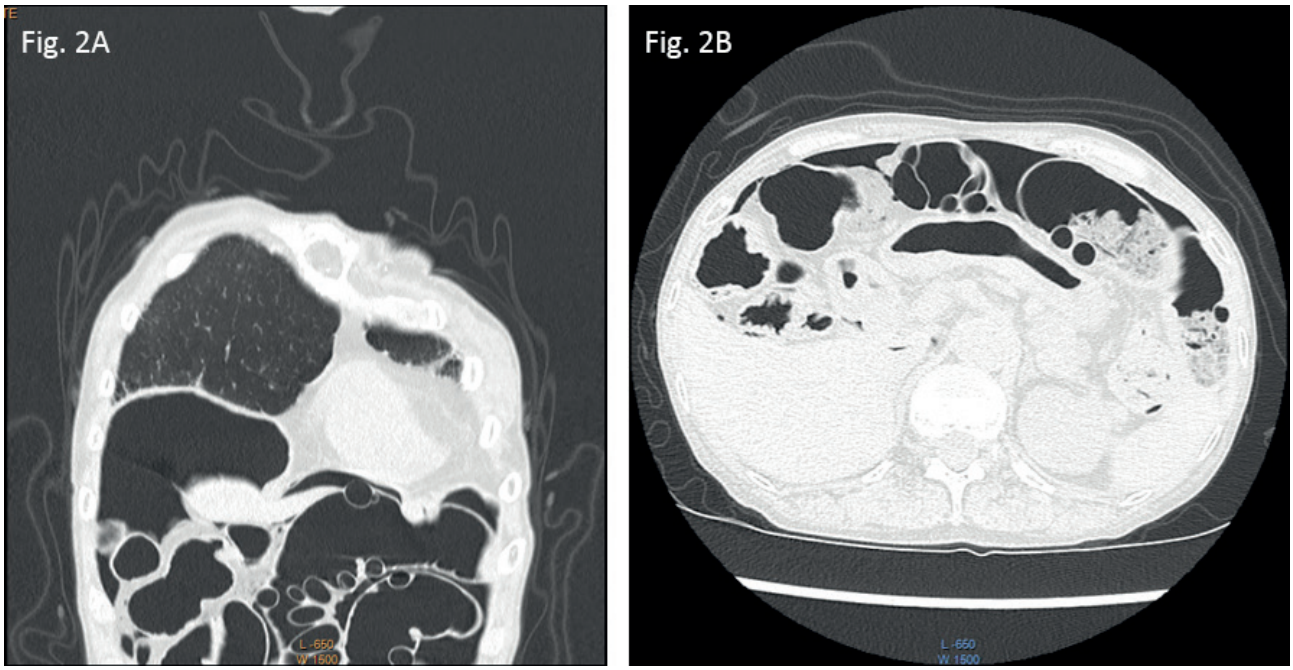


Figure 2A, B. Abdominal CT scan reveals the exacerbation of pneumoperitoneum, dilated intestinal canals, and air-filled cysts of the walls.

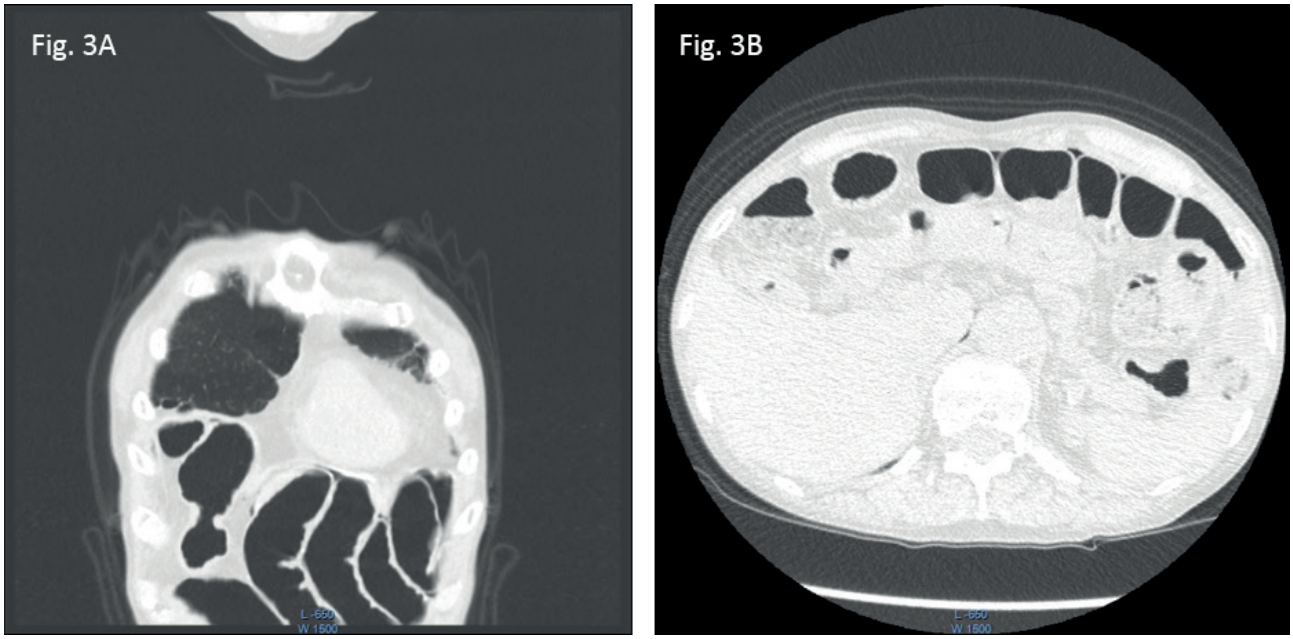


Figure 3A, B. Abdominal CT scan reveals the improvement of pneumoperitoneum, dilated intestinal canals, and air-filled cysts of the walls.

that treatments with cisapride, diet, and oxygen inhalation were efficacious [5,6]. PI associated with SjS is a rare complication, and the etiologies remain unclear. Therefore, more research is required to elucidate the etiologies, as well as prompt and precise diagnosis and efficacious treatments.

Conflict of interest

The authors declare that there is no conflict of interest.

REFERENCES

[1] Suzuki E, Kanno T, Hazama M, Kobayashi H, Watanabe H, Ohira H. Four cases of pneumatosis cystoides intestinalis complicated by connective tissue diseases. Intern Med. 2017;56(9):1101-6. doi: 10.2169/internalmedicine.56.7877. Epub 2017 May 1. PMID: 28458320; PMCID: PMC5478575.

[2] Pear BL. Pneumatosis intestinalis: a review. Radiology 1998;207:13–9.

[3] Sagara A, Kitagawa K, Furuichi K, Kitajima S, Toyama T, Okumura T, Hara A, Sakai Y, Kaneko S, Wada T. Three cases of pneumatosis intestinalis presenting in autoimmune diseases. Mod Rheumatol. 2012;22(4):610-15. doi: 10.1007/s10165-011-0551-3. Epub 2011 Nov 9. PMID: 22068684.

[4] Martis N, Baque-Juston M, Buscot M, Marquette CH, Leroy S. An unexpected discovery complicating sicca syndrome. Eur Respir Rev. 2014 Sep;23(133):397-8. doi: 10.1183/09059180.00000114. PMID: 25176978.

[5] Vandergheynst F, Van Gansbeke D, Steinfeld S. Primary Sjögren's syndrome associated with pneumatosis cystoides coli. Eur J Intern Med. 2006;17(1):57-8. doi: 10.1016/j.ejim.2005.06.002. PMID: 16378889.

[6] Ikeda H, Toyonaga A, Sakai E, Suga H, Kawano H, Fujita M, et al. A case of pneumatosis cystoides coli associated with Sjogren's syndrome. Dig Endosc 1996;8(4):302-5. doi: 10.1111/j.1443-1661.1996.tb00443.x.

[7] Mizoguchi F, Nanki T, Miyasaka N. Pneumatosis cystoides intestinalis following lupus enteritis and peritonitis. Intern Med. 2008;47:1267-71. doi: 10.2169/internalmedicine.47.0748. Epub 2008 Jul 1. PMID: 18591854.